



Dupuytren contracture

Dupuytren contracture is a deformity of the hand in which the joints of one or more fingers can become permanently bent in a flexed position. Permanently bent joints are called contractures. The condition most often occurs in men older than age 50. In women, it is four times less common, and also tends to appear later and be less severe. However, Dupuytren contracture can occur at any time of life, including childhood. The disorder can make it more difficult for affected individuals to perform manual tasks such as preparing food, writing, or playing musical instruments.

In about half of cases, Dupuytren contracture occurs in only one hand, affecting the right hand twice as often as the left. Which hand is affected does not seem to be related to whether the person is right-handed or left-handed.

Dupuytren contracture results from shortening and thickening of bands of fibrous tissue under the skin of the palm (palmar fascia). Fascia is a type of connective tissue, which supports the body's muscles, joints, organs, and skin and provides strength and flexibility to structures throughout the body.

In Dupuytren contracture the thickening of the fascia typically first appears as one or more small hard nodules that can be seen and felt under the skin of the palm. In some affected individuals the nodules remain the only sign of the disorder, and occasionally even go away without treatment, but in most cases the condition gradually gets worse. Over months or years, the abnormal fibrous tissue gets shorter and thicker, developing into tight bands of tissue called cords. These cords gradually draw the affected fingers downward so that they curl toward the palm. As the condition gets worse, it becomes difficult or impossible to extend the affected fingers, resulting in the contracture associated with this disorder. The ring finger is most often involved, followed by the little, middle, and index fingers. Occasionally the thumb is involved.

Dupuytren contracture is usually not painful, but in some cases people with this condition experience uncomfortable joint inflammation or sensations of burning or itching. Pressure or tension may also be experienced, especially when attempting to straighten affected joints.

People with Dupuytren contracture are at increased risk of developing other disorders in which similar connective tissue abnormalities affect other parts of the body. These include Garrod pads, which are nodules that develop on the knuckles; Ledderhose disease, also called plantar fibromatosis, in which contractures affect the foot; and, in males, Peyronie disease, which causes abnormal curvature of the penis.

Frequency

Dupuytren contracture occurs in about 5 percent of people in the United States. It is common in northern Europeans; 30 percent of Norwegian men over age 60 develop the disorder. Studies suggest that a genetic predisposition to develop this disorder may have been spread through northern Europe and Britain by the Vikings. Dupuytren contracture is less common in non-European populations.

Genetic Changes

While the cause of Dupuytren contracture is unknown, changes in any of several genes are thought to affect the risk of developing this disorder. Some of these genes are involved in a biological process called the Wnt signaling pathway. This pathway promotes the growth and division (proliferation) of cells and is involved in the determination of the specialized functions a cell will have (differentiation).

Abnormal proliferation and differentiation of connective tissue cells called fibroblasts are important in the development of Dupuytren contracture. The palmar fascia of people with this disorder has an excess of myofibroblasts, which are a type of fibroblast containing protein strands called myofibrils. Myofibrils normally form the basic unit of muscle fibers, allowing them to contract. The increased number of myofibroblasts in this disorder cause abnormal contraction of the fascia and produce excess amounts of a connective tissue protein called type III collagen. The combination of abnormal contraction and excess type III collagen likely results in the shortening and thickening of fascia that occurs in Dupuytren contracture. However, it is unknown how changes in genes that affect the Wnt signaling pathway are related to these abnormalities and how they contribute to the risk of developing this disorder.

Other risk factors for developing Dupuytren contracture may include smoking; heavy alcohol use; liver disease; diabetes; high cholesterol; thyroid problems; certain medications, such as those used to treat epilepsy (anticonvulsants); and previous injury to the hand. The higher prevalence of the disorder in males than in females is thought to be due to sensitivity of the fascia to male sex hormones (androgens).

Inheritance Pattern

Dupuytren contracture is usually passed down in families and is the most common inherited disorder of connective tissue. The inheritance pattern is often unclear. Some people who inherit gene changes associated with Dupuytren contracture never develop the condition.

In some families, Dupuytren contracture appears to be inherited in an autosomal dominant pattern, which means one copy of an altered gene in each cell is sufficient to cause the disorder. In these cases, an affected person usually has one parent with the condition.

In other families, the condition appears to be inherited in a mitochondrial pattern, which is also known as maternal inheritance. This pattern of inheritance applies to genes contained in mitochondrial DNA (mtDNA), a form of DNA found in energy-producing structures within cells (mitochondria). Because egg cells, but not sperm cells, contribute mitochondria to the developing embryo, children can inherit disorders resulting from mtDNA mutations only from their mother. These disorders can appear in every generation of a family and can affect both males and females, but fathers do not pass traits associated with changes in mtDNA to their children.

In some cases Dupuytren contracture is not inherited and occurs in people with no history of the condition in their family. These sporadic cases tend to begin later and be less severe than familial cases.

Other Names for This Condition

- contraction of palmar fascia
- Dupuytren's contracture
- Dupuytren's disease
- familial palmar fibromatosis
- palmar fascial fibromatosis
- palmar fibromas

Diagnosis & Management

Genetic Testing

- Genetic Testing Registry: Dupuytren's disease of palm, with contracture
<https://www.ncbi.nlm.nih.gov/gtr/conditions/C0013312/>

Other Diagnosis and Management Resources

- American Society for Surgery of the Hand
<http://www.asssh.org/handcare/Anatomy/Details-Page/ArticleID/27961/Dupuytren-Disease>
- National Data Bank for Rheumatic Diseases: Joining NDB Research for Dupuytren
<https://www.arthritis-research.org/join-research/joining-ndb-research-dupuytren>
- NHS Choices (UK)
<http://www.nhs.uk/conditions/dupuytren-s-contracture/pages/introduction.aspx>
- The Cochrane Collaboration: Surgery for Dupuytren's Disease of the Fingers
http://www.cochrane.org/CD010143/MUSKEL_surgery-dupuytren-s-disease-fingers

General Information from MedlinePlus

- Diagnostic Tests
<https://medlineplus.gov/diagnostictests.html>
- Drug Therapy
<https://medlineplus.gov/drugtherapy.html>
- Genetic Counseling
<https://medlineplus.gov/geneticcounseling.html>
- Palliative Care
<https://medlineplus.gov/palliativecare.html>
- Surgery and Rehabilitation
<https://medlineplus.gov/surgeryandrehabilitation.html>

Additional Information & Resources

MedlinePlus

- Encyclopedia: Contracture Deformity
<https://medlineplus.gov/ency/article/003185.htm>
- Encyclopedia: Dupuytren Contracture
<https://medlineplus.gov/ency/article/001233.htm>
- Encyclopedia: Finger Pain
<https://medlineplus.gov/ency/article/003248.htm>
- Health Topic: Connective Tissue Disorders
<https://medlineplus.gov/connectivetissuedisorders.html>
- Health Topic: Finger Injuries and Disorders
<https://medlineplus.gov/fingerinjuriesanddisorders.html>
- Health Topic: Hand Injuries and Disorders
<https://medlineplus.gov/handinjuriesanddisorders.html>

Genetic and Rare Diseases Information Center

- Familial Dupuytren contracture
<https://rarediseases.info.nih.gov/diseases/12165/familial-dupuytren-contracture>

Educational Resources

- Canadian Centre for Occupational Health and Safety: Dupuytren's Contracture
<https://www.ccohs.ca/oshanswers/diseases/dupuytre.html>
- MalaCards: familial dupuytren contracture
https://www.malacards.org/card/familial_dupuytren_contracture

- Merck Manual Consumer Version: Dupuytren Contracture
<http://www.merckmanuals.com/home/bone,-joint,-and-muscle-disorders/hand-disorders/dupuytren-contracture>
- Orphanet: Familial Dupuytren contracture
http://www.orpha.net/consor/cgi-bin/OC_Exp.php?Lng=EN&Expert=79142

Patient Support and Advocacy Resources

- American Academy of Orthopaedic Surgeons
<http://orthoinfo.aaos.org/topic.cfm?topic=a00008>
- Dupuytren Foundation
<https://dupuytren.org/>
- International Dupuytren Society
<http://www.dupuytren-online.info/>
- National Organization for Rare Disorders
<https://rarediseases.org/rare-diseases/dupuytren-contracture/>
- The British Dupuytren's Society
<http://dupuytren-society.org.uk/>

ClinicalTrials.gov

- ClinicalTrials.gov
<https://clinicaltrials.gov/ct2/results?cond=%22Dupuytren+contracture%22>

Scientific Articles on PubMed

- PubMed
<https://www.ncbi.nlm.nih.gov/pubmed?term=%28Dupuytren+Contracture%5BTIAB%5D%29+AND+english%5Bla%5D+AND+human%5Bmh%5D+AND+%22last+1800+days%22%5Bdp%5D>

OMIM

- DUPUYTREN CONTRACTURE
<http://omim.org/entry/126900>

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